A stitch in time saves sight: Craniopharyngioma

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Craniopharyngiomas are rare, relatively benign neoplasms that typically arise in the sellar or parasellar region, which can be solid, cystic, or calcified. We are reporting a case of an 11-year-old female child who presented with complaints of sudden onset rapid diminution of vision in the right eye more than the left eye for 2 months, for distant and near vision, with normal visual fields. The patient was diagnosed as having craniopharyngioma based on magnetic resonance imaging. Surgery was done without delay, restoring her vision. Histopathology of tumor confirmed the diagnosis. This case highlights the fact that immediate neuroimaging in children presenting with sudden onset, rapidly progressive vision loss helps in early diagnosis and management of craniopharyngioma and to retrieve lost vision.

Key words: Craniopharyngioma, Magnetic resonance imaging, Surgical management, Visual loss

Craniopharyngiomas are rare, relatively benign neoplasms that typically arise in the sellar/parasellar region. They are derived from embryonic cellular remnants of the Rathke’s pouch found along the path of the primitive adenohypophyseal and craniopharyngeal duct. The prevalence of craniopharyngioma is 0.13–2 per 100,000 [1]. It occurs due to a defect in Wnt signaling pathway reactivation due to B catenin gene mutation. Craniopharyngiomas are primarily suprasellar tumors (75%) while a small intrasellar component is present in 20–25% of cases.

Occasionally, craniopharyngiomas appear as intraventricular, homogeneous, soft-tissue masses without calcification (papillary subtype). The third ventricle is a particularly common location. Rare or ectopic locations reported include nasopharynx, posterior fossa, and extension down the cervical spine. The tumor has a bimodal distribution. The first peak occurs between the ages of 5 and 15 years and a second smaller peak occurs in adults aged over 40 years [1]. Clinical presentation is varied on account of the variable location and size of the tumor. Presenting complaints include headache, nausea, vomiting, visual symptoms, and endocrine dysfunction.

CASE REPORT

An 11-year-old female child born of non-consanguineous marriage with normal birth history and developmental milestones, presented with complaints of sudden onset, progressive rapid diminution of vision in the right eye more than the left eye for 2 months and intermittent headache. Initially, the child complained of blurred vision for distant vision. There was no history of specific visual field loss. Two months later, she could not identify coins or see numbers on the mobile. Family history was not significant.

On general examination, the patient’s vitals were normal. On neurological examination, higher functions were normal. Examination of the second cranial nerve showed visual acuity of finger counting at 2 feet and 3 feet in the right and left eye, respectively. However, visual fields were normal on the ophthalmological examination. Other cranial nerves were normal. There was no motor or sensory deficit or cerebellar signs.

Multiplanar magnetic resonance imaging (MRI) brain showed thin-walled predominantly cystic sellar-suprasellar lesion, measuring 3 × 3.6 × 4 cm and pituitary gland was not seen. These features were suggestive of craniopharyngioma (Fig. 1). Hormonal assays showed low cortisol level - one microgram/dl (normal range = 1–22 microgram/dl) and thyroid-stimulating hormone (TSH) 5.8 Mu/L (normal range = 0.55–5.31 Mu/L).

The patient was treated immediately. Surgical management was done with total excision of the tumor and the excised specimen sent for histopathology. Histopathology of the tumor revealed solid and cystic areas with cells arranged in large islands and sheets with multiple foci of calcifications suggestive of adamantinomatous craniopharyngioma WHO Grade 1. The patient was also started on steroids 1 mg/kg/day in view of low cortisol and oral thyroxine 25 microgram once daily in the morning (empty stomach) in view of hypothyroidism. Postoperatively, vision recovered completely.

DISCUSSION

Craniopharyngioma is a relatively rare benign slow-growing tumor which has a prevalence of 0.13–2 per 100,000 [1]. The first description of a craniopharyngioma was given in 1857 by Zenker.
The term craniopharyngioma was introduced by Cushing in 1932. Thereafter, a detailed study was done and its histological subtypes were identified. The diagnosis of this tumor is based on clinical presentation and radiological findings.

Craniopharyngioma presents with visual disturbances, increased intracranial pressure, neurobehavioral and endocrine problems. In a study by Wan et al., the most common presenting features were headache (76%), nausea or vomiting (32%), and vision loss (31%). Our patient presented with visual disturbance and intermittent headache without any nausea or vomiting. Various reports have described neurological disturbances such as headache, vision loss, and visual field defects, along with manifestations of endocrine deficiency such as growth retardation and delayed puberty to be the most common presenting symptoms of craniopharyngiomas. Although our case presented with a diminution of vision, there was no visual field defect. These tumors can stretch the diaphragm of the sella and cause headaches. Obstruction of the cerebral aqueduct and the foramen of Monro may also lead to hydrocephalus, which makes a shunt necessary. Hydrocephalus was not seen in our patient.

At diagnosis, endocrine dysfunction is found in up to 80% of patients. Reduced growth hormone secretion is the most frequent endocrinopathy and can be present in up to 75% of patients. This is followed by follicle-stimulating hormone/luteinizing hormone deficiency, which can be seen in 40% of patients and then adrenocorticotropic hormone and TSH deficiency in 25%. Our patient had low cortisol levels and hypothyroidism which is a less commonly seen endocrinopathy in craniopharyngioma. A study by DeVile et al. states that hypoadrenalism and associated hypoglycemia contribute to morbidity and mortality in children with craniopharyngioma with endocrine dysfunction.

The diagnosis on MRI is mainly based on the three characteristic components of the tumor: Cystic, solid, and calcified. The cystic component constitutes the most important part of the tumor and shows variable signal depending on the chemical-physical
Craniopharyngioma remains a challenging tumor because despite its benign histological appearance, it is often associated with unfavorable and occasional disastrous sequelae. Hence, a high index of suspicion in children presenting with visual symptoms can lead to early diagnosis and treatment. Immediate neuroimaging and surgery in our patient with craniopharyngioma, who presented with sudden onset and rapidly progressive vision loss, resulted in complete retrieval of lost vision. Early diagnosis with neuroimaging and immediate surgical treatment is essential for a good outcome. However, endocrine abnormalities require treatment and long-term monitoring even after surgery. All children with craniopharyngioma should be managed and followed up by a multidisciplinary team.

REFERENCES


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